Uncommon vascular thoracic and abdominal anomalies detected with MDCT and 1.5T MR in pediatric population.

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Learning objectives

To review the uncommon congenital vascular thoracic and abdominal anomalies in pediatric patients, and to describe the CT and MR angiography protocol used in our institution in pediatric population.

We present CT and MR findings cases of uncommon congenital vascular thoracic and abdominal anomalies (Aortic coarctation, Left superior vena cava persistent, Right aortic arch with Kommerel's diverticulum of left subclavian artery, Pulmonary sequestration, Congenital absence of portal vein, Acute thrombosis of portal system, portal-venous shunt intra and extrahepatic).

Background

Uncommon vascular anomalies in pediatric population represent in most cases incidental findings. Their detection is critical because these anomalies can be responsible of serious complication during surgery.

16MDCT, 64MDCT and 1.5T MR studies, using an angiography protocol, MPR, MIP and VR analysis are fundamental to detect these anomalies.

Imaging findings OR Procedure details

MDCT technique: Our technique and protocol are showed in Figures 1 and 2.

MR technique: Our technique and protocol are showed in Figures 3 and 4.

Kawasaki: Kawasaki disease (KD) is an acute multisystemic vasculitis occurring predominantly in children and rarely in adults. It is characterized by fever, bilateral nonexudative conjunctivitis, erythema of the lips and oral mucosa, changes in the extremities, rash, and cervical lymphadenopathy.

During the acute phase 10-25% of patients develop coronary ectasia or coronary artery aneurysms (CAA).

Most CAA are small to medium in size
The so-called "giant" aneurysms exceed 8 mm in diameter. In the long term these pathological changes may lead to thrombotic vessel occlusion, coronary stenosis or premature arteriosclerosis.

The CCA may lead to ischemic heart disease or sudden death. For this reason the follow-up coronary artery imaging is essential.

**Figure 5.**

**Aortic coarctation:** Aortic coarctation, described by Morgagni in 1760, involves aortic narrowing in the region of the ligamentum arteriosum just distal to the left subclavian artery. It has a male predominance (male-to-female ratio, 1.5:1). The stenotic segment frequently develops in a juxtaductal location but may show extension into the aortic arch and isthmus.

Aortic coarctation occurs as a solitary lesion in 82% of cases but may be associated with other pathologies as bicuspid aortic valve (almost 30% of cases), Turner syndrome, intracranial aneurysms (10% of cases), VSD and atrial septal and valve defect, and with isthmic hypoplasia, aortic arch hypoplasia or both. Figure 6.

**Takayasu:** Takayasu arteritis is a primary large-vessel arteritis of unknown origin that affects the aorta and its main branches. 80%-90% of patients with Takayasu arteritis are female and in the second or third decade of life. CT angiography clearly depicts all aortic luminal changes, including stenosis, dilatation, and aneurysm. Furthermore, axial images demonstrate circumferential wall thickening of the aorta and its involved branches, thus allowing detection of Takayasu arteritis in the early systemic phase, when inflammation or thickening of the vessel wall may already be seen in the absence of luminal abnormalities. Figures 7,8,9.

**Left superior vena cava persistent:** A persistent left superior vena cava (PLSVC) is the most common thoracic venous anomaly occurring in approximately 0.5% of the normal population. It is a persistent remnant of a vessel that is present as a counterpart of normal right-sided superior vena cava (SVC) in early embryological development but normally disappears later. It may be associated with a missing right superior vena cava or other cardiac anomalies.

If it is not associated with other congenital cardiac anomalies, it is usually asymptomatic and hemodynamically insignificant. In these cases, PLSCV is an incidental finding during diagnostic and therapeutic procedures. Although it can be easily diagnosed by the characteristic chest radiographic appearance of a pulmonary artery catheter (PAC) passed through it after being inserted into the left subclavian or jugular vein, its diagnosis is usually missed by the presence of normal (right) SVC and the passage of the catheter on the right side. Its diagnosis can be confirmed by many noninvasive and invasive
tests, or it is incidentally diagnosed at thoracic surgery or autopsy. However, PLSVC has important clinical implications in certain situations. Figure 10.

**Kommerel’s diverticulum:** Kommerell diverticulum (KD) represents the remnant of the left fourth arch and is situated at the point of merger between the aortic arch and the proximal descending thoracic aorta.

KD is an aortic anomaly characterized by a left aortic arch and an aberrant right subclavian artery arising as the last branch of the aortic arch and courses from the proximal descending aorta to the right arm. The aberrant right subclavian artery cross behind the esophagus and for this reason can, but do not always, cause symptoms of tracheal or esophageal compression.

Kommerell diverticulum may be present either with a left aortic arch, but KD is most frequently present in cases of right-sided aortic arch with an aberrant left subclavian artery. In this vascular anomaly, the left subclavian artery arises from KD and passes obliquely upward, behind the esophagus, toward the left arm. Figure 11.

**Pulmonary sequestration:** PS is characterized by a portion of lung that is segregated from the remainder of the normal lung and receive a systemic arterial supply, usually from the thoracic or abdominal aorta. Two types of sequestration are known: intralobar and extralobar. The intralobar sequestration accounts almost the 75% of bronchopulmonary sequestration. Most patients are asymptomatic, until an acute respiratory infection develops. The most common radiographic presentation is a homogeneous opacity in the posterior basal segment of a lower lobe (more frequently the left). The diagnosis is based on the demonstration of the anomalous systemic supply to the sequestered lobe.

The Extralobar sequestration may be associated with other congenital systemic anomalies, such as congenital diaphragmatic hernia, cardiac abnormalities, pulmonary hypoplasia, or foregut duplication cysts. It may be located below the diaphragm and may mimic a neuroblastoma or adrenal hemorrhage.

The extralobar form has its own pleural investment and systemic venous drainage, whereas the intralobar form shares the pleural investment with the normal lung and usually (but not invariably) drains into the pulmonary venous system. It is believed to arise from a supernumerary lung bud caudad to the normal lung bud. If the lung bud arises before the development of the pleura, it is invested with adjacent lung and becomes an intralobar sequestration. If the lung bud arises after pleura formation, it grows separately and acquires its own pleural covering. Figure 12.

**Congenital absence of portal vein:** Congenital absence of the portal vein (CAPV) is a rare anomaly in which the intestinal and the splenic venous drainage bypass the liver and drain into systemic veins through various venous shunts. Two types of shunt exist:
either the liver is not perfused with portal blood because of a complete shunt (Type I) or
the liver is perfused with portal blood due to the presence of a partial shunt (Type II).

Type I CAVP is a rare anomaly, frequently associated with other congenital anomalies, more frequent in females (74%), and characterized by a drainage into the systemic vein bypassing the liver. CAVP, usually asymptomatic, can cause minimal abnormalities of liver enzymes. In adulthood, CAVP is often complicated by hepatic encephalopathy, hepatopulmonary syndrome or hepatic tumors.

Two forms of CAVP Type I are known.

Ia: Superior mesenteric vein (SMV) and splenic vein (SpV) drain separately into inferior vena cava (IVC).

Ib: SMV and SpV form a common trunk before draining into the IVC. Figures 13, 14.

Absence of the inferior vena cava: The absence of the inferior vena cava is a rare congenital anomaly characterized by a developmental defect of the IVC and collateral circulation. The prevalence of congenital absence or interrupted IVC has been reported at 0.15%-3% and represents a modest risk for deep-venous thrombosis. When associated with genetic polymorphism or other hypercoagulable states, the risk can increase several times. In patients with congenital absence of the IVC, alternative routes for venous return form; these alternative routes can include hemi-azygous, vertebrolumbar, anterior abdominal wall, and transumbilical portocaval collaterals. Of interest, if these collateral vessel systems are well formed, patients tend to be asymptomatic. Figure 15.

Portal-venous and veno-venous intrahepatic shunt: Intrahepatic portosystemic shunts are infrequent in children. Park classified them as being 1 of 4 types: I, a single large vessel connecting the right portal vein to the inferior vena cava;

II, a peripheral localized shunt in which single or multiple communications are present between peripheral branches of the portal and hepatic veins in 1 hepatic segment;

III, an aneurysm connecting the peripheral portal and hepatic veins;

IV, diffuse communication between the peripheral portal and hepatic veins in both lobes.

We found that patients with biliary atresia frequently showed portal-venous and/or veno-venous communication in the peripheral areas of the liver. Veno-venous shunts are usually detected in the portal-venous phase near the liver surface in the axial plane and confirmed in 3D reconstructions. Interestingly, one of the paradigmatic liver diseases of adulthood with presinusoidal portal hypertension, non-cirrhotic portal hypertension (which includes idiopathic portal hypertension, hepatoportal sclerosis and nodular regenerative hyperplasia) is the situation in which veno-venous shunts were first described; furthermore veno-venous shunts can also be present in hepatic
schistosomiasis, another disease characterised by a presinusoidal haemodynamic pattern of portal hypertension. As in the case of Idiopathic Portal Hypertension, multiple occlusions of small-sized intrahepatic portal branches may be responsible for a gradual loss of parenchymal cells near the liver surface, thus promoting the production of shunting. Figure 16,17.

**Gastrointestinal angiodysplasia:** Gastrointestinal angiodysplasia is an important vascular lesion responsible for approximately 6% of cases of lower gastrointestinal hemorrhage. Patients with colonic angiodysplasia may present with hematochezia, melena, positive results on the Hemoccult Fecal Occult Blood Test (Beckman Coulter), or iron deficiency anemia. The exact mechanism underlying the development of angiodysplasia is unclear. The most prominent hypothesis is related to the degenerative process of small blood vessels that is associated with aging. Arterio-venous fistulae (AVF) of the colon, of various etiologies, are rare in children, and more frequently affect the left hemicolon. These malformations can have varied symptoms, but lower gastrointestinal bleeding and severe anemia are the most common. AVF that involve the portal system can cause extrahepatic portal hypertension in non-cirrhotic livers, as well as gastric variceal bleeding.

These lesions can be suspected if the alteration of the Doppler waveform in the portal system is present, and can later be confirmed with Angio-MDCT.

Portal hypertension and gastro-intestinal bleeding secondary to AVF are surgically treatable with ligation of the feeding artery, or resection of the fistula. Figure 18.

**Thrombosis of portal system:** Portal vein thrombosis (PVT) is a major cause of portal hypertension in children and adolescents. PVT may be related to neonatal events associated with the physiologic process of closure of the umbilical vein and ductus venosus or to developmental defects. In a considerable number of cases, however, the etiology remains not clearly defined. Genetic abnormalities affecting the physiologic anticoagulant system, such as hereditary deficiency of protein C (PC), protein S (PS), and antithrombin (AT), have been well established as risk factors of venous thrombosis in adults. The recently described factor V Leiden (FVL), methylenetetrahydrofolate reductase (MTHFR) C677T, and prothrombin (PTHR) G20210A mutations have also been reported as risk factors in adult PVT patients.

The initial clinical manifestation is characterized either by episodes of upper gastrointestinal bleeding or by splenomegaly on routine clinical examination. The major complications include upper gastrointestinal bleeding, hypersplenism secondary to splenomegaly, growth retardation, and portal biliopathy. The diagnosis is made by abdominal Doppler ultrasonography. Figure 19.
The procedure is performed under monitored anaesthesia care, with intravenous administration of midazolam (0.15-0.2 mg/kg), ketamine (1.5-3 mg/kg) or propofol (0.5-1 mg/kg), with spontaneous respiration or general anaesthesia. No high density oral contrast medium is administered. The patient is positioned supine with feet first on the imaging table. After acquisition of anteroposterior and mediolateral digital scout radiographs the images of the region of interest is acquired, after intravenous contrast medium administration, in the cranial-caudal direction; tube current is modified according to body weight only in children over 27 Kilos. The contrast medium used is non-ionic isomolar 320 mg l/ml, 1.5 ml/Kg at a flow rate depending on the weight of the patient and on the gauge of the intravenous access (<5Kg, 0.8 ml/sec; 5-10Kg, 1.2 ml/sec; 10-15Kg, 1.5 ml/sec; 15-20Kg, 2.5 ml/sec; 20-30Kg, 3 ml/sec; 30-50Kg, 3.5 ml/sec; 50-60Kg, 4 ml/sec). Contrast medium is administered by means of a mechanical power injector and followed by a saline flush of 0.3 ml/Kg. Two phases are acquired. Bolus tracking technique is used: the region of interest (ROI) of the bolus-tracking technique is placed in the aorta. After contrast injection, when the ROI reach 100 UH, the arterial acquisition start craniocaudally after a 5-second post threshold delay. The inherent 5-second delay in the bolus-tracking technique is necessary to move the scan table to the start of the scan. The time of acquisition of the portal-venous phase is obtained 50 sec after starting the contrast medium injection. All phases are acquired without breath-holding. A console for post-processing is used to perform Multi Plane Reconstruction (MPR), Multi Intensity Projection (MIP) and Volume Rendering (VR) vascular reconstruction.

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**Fig. 2**
The procedure is performed with imaging acquisition in breath holding.
Supine position with feet first on the imaging table.
8ch phase array body coil is used for a good quality of the angiography images.
The contrast medium used is a non-ioninc gadolinium based (0.5 M). 1ml/kg at flow rate
(from 0.8 to 1 ml/sec) depend on the weight of the patient and on the gauge of intravenous
access.

After acquisition of localizzazione images on three planes (Axial, Sagittal and Coronal),
the procedure divides in two phases:

1) 2D FIESTA (Fast Imaging Employing Steady state Acquisition) Gradient Echo sequences on
Axial, Coronal and oblique plane. FIESTA provides images of fluid filled structures with very
short acquisition times. These images can also acquired with respiratory trigger without breath
holding of the patient.

2) 3D CE-MRA (Contrast Enhancement Magnetic Resonance Angiography) spoiled gradient
echo pulse sequences. Contrast enhanced MRA is performed with a short TR to have low signal
(due to the longer T1) from the stationary tissue, short scan time to facilitate breath hold
imaging, short TE to minimize T2* effects and a bolus injection of a sufficient dose of a
gadolinium chelate (Prohance).

Fig. 3
3D imaging volume preferred for thoracic MRA is oblique along a imaginary axis including ascendent and descendent aorta. Before acquisition of the contrast images, it is necessary acquired a mask phase. The arterial phase is acquired when the aorta is full of contrast medium (Fluoro Trigger technique). After arterial phase, other two phases are acquired.
A console for post-processing (Advantage Windows 4.1 and 4.2 GE) is used to perform Multi Plane Reconstruction (MPR), Multi Intensity Projection (MIP), and Volume Rendering (VR) vascular reconstruction.

2D FIESTA GE sequence parameter on Axial and Sagittal Planes
Matrix 200 (Phase) X 160 (Frequency), Nex 1, Phase Fov 1, Frequency/Direction A/P, Slice Thickness 4.0 mm, Overlap spacing, TE Minimum Full, Flip Angle 75, Bandwith 80.83 KHz, Imaging Options (Extending Dynamic Range, Asset, Flow Compensation).

2D FIESTA GE sequence parameter on Coronal Plane
Matrix 256 (Phase) X 192 (Frequency), Nex 1, Phase Fov 1, Frequency/Direction R/L, Slice Thickness 4.0 mm, Overlap spacing, TE Minimum Full, Flip Angle 60, Bandwith 83.33 KHz, Imaging Options (Extending Dynamic Range, Flow Compensation).

3D CE-MRA spoiled gradient echo pulse sequence parameters
Matrix 256 (Phase) X 192 (Frequency), Nex 0.5, Phase Fov 1, Frequency/Direction A/P, Slice Thickness 1.2-2 mm, Overlap spacing, TE Minimum Full, Flip Angle 20, Bandwith 100 KHz, Imaging Options (Asset, Eliptic Centric, Turbo Mode 1).

Fig. 4
**Fig. 5:** Kawasaki disease in 8 year old female child. MDCT MIP reconstruction show a giant aneurysm (10 mm) of the left coronary artery (LCA). Of note is the presence of calcification in the LCA.
**Fig. 6:** Aortic Coarctation in 5 year old female child. Volume rendering reconstruction of AngioMRI shows an irregular and long stenosis of the isthmus and proximal descending aorta.

![Aortic Coarctation](image)

**Fig. 7:** Takayasu disease in 9 year old female child. MDCT Volume Rendering reconstruction shows an irregular stenosis of the left subclavian artery.

![Takayasu Disease](image)
**Fig. 8:** Takayasu disease in 9 year old female child. MDCT Multi plane reconstruction (MPR) in coronal image shows an irregular stenosis of the sub/renal aorta caused by circumferential wall thickening.
**Fig. 9:** Same patient of figure 8. MDCT Volume Rendering reconstruction shows also a mild stenosis of the origin of the right renal artery, and confirms a narrowing of the aorta.

**Fig. 10:** A 9-year-old girl with end-stage kidney disease secondary to right renal agenesis. MDCT Volume rendering reconstruction shows left innominate vein (in yellow, comprised of left internal jugular vein and left subclavian vein) and superior vena cava (in red). The superior vena cava drains the right innominate vein and the left external jugular vein. A thin anastomotic branch between the left innominate vein and the superior vena cava can be seen.
**Fig. 11:** 2 month old male baby. MDCT volume rendering reconstruction shows right (white arrows) and left (red arrows) aberrant subclavian arteries arising at the point of merger between the aortic arch and the proximal descending thoracic aorta.

**Fig. 12:** Intralobar pulmonary sequestration in 21 month old male baby. MDCT volume rendering reconstruction (A) and multi intensity projection (B) show aberrant arterial vessel arising from the aorta, that supply the basal segment of the left inferior lobe. Of note is the presence of normal venous drainage of the sequestered parenchyma.
Fig. 13: CAVP in 9 month old female baby. MDCT VR-reconstruction shows a common trunk formed by SMV and SpV draining into IVC, and multiple intrahepatic venous-venous shunts.
Fig. 14: CAVP in 11 month old female baby. MDCT VR-reconstruction shows a common trunk formed by SMV, SpV and left renal vein draining into IVC.
Fig. 15: Absence of Inferior vena cava (IVC) in 4 year old female child; status post OLTx. Multi plane reconstruction images show absence of IVC (white arrows) associated with hypertrophy of lumbar veins, and hypertrophic azygos and emiazygos venous system (green arrows).
**Fig. 16:** 15 month old boy with biliary atresia. MDCT Volume Rendering reconstruction shows bilobar multiple portal-venous shunts (arrows).
**Fig. 17:** 13 month old girl with biliary atresia. MIP reconstruction shows multiple tortuous veno-venous shunts in the left lobe (arrow). Of note, portal hypertension signs represented by large para-oesophageal varices (V).
Fig. 18: 14 year old male with bloody diarrhea, vomiting, severe abdominal pain, and fever. MDCT-Volume-Rendering confirms large Arterio-Venous Fistula in the left colon (between inferior mesenteric artery, and inferior mesenteric vein); coexist AVF in the duodenum (superior mesenteric branch).
**Fig. 19:** A 12 year-old child with MTHFR heterozygosis. Multi plane reconstruction shows a large thrombosis of portal system.
Conclusion

MDCT, and 1.5T MR, using a angiography protocol in pediatric patient is helpful in detection of uncommon congenital vascular thoracic and abdominal anomalies.

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